

Hepatic rupture in HELLP syndrome. Review of surgical treatment

Ruptura hepática en el síndrome de HELLP. Revisión del tratamiento quirúrgico

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ABSTRACT

Introduction: Hepatic rupture in HELLP syndrome is a complication that is fatal for the mother-child pair in up to 80% of cases. Multiple therapeutic options have been described, such as hepatic packing, segmental resections, and even liver transplantation for severe cases. **Clinical case:** A cesarean section was performed in a 32-year-old woman with HELLP syndrome and a hepatic subcapsular hematoma was found in the left lobe. Initially hepatic packing and ligation of the common hepatic artery were carried out, and later a left hepatectomy was done as definitive management. **Discussion and conclusions:** The reported case was successfully treated by various surgical methods. Early recognition of this complication, aggressive surgical management and multidisciplinary support are the basis for reducing morbidity and mortality.

RESUMEN

Introducción: La ruptura hepática en síndrome de HELLP es una complicación que resulta mortal para el binomio madre-hijo hasta en un 80% de los casos. Múltiples opciones terapéuticas se han descrito, tales como empaquetamiento hepático, resecciones segmentarias, e incluso trasplante hepático para casos severos. **Caso clínico:** Mujer de 32 años con síndrome de HELLP, se realiza cesárea encontrándose hematoma subcapsular hepático en lóbulo izquierdo, inicialmente se realizó empaquetamiento hepático y ligadura de la arteria hepática común, y hepatectomía izquierda como manejo definitivo. **Discusión y conclusiones:** El caso reportado fue tratado exitosamente mediante diversos métodos quirúrgicos. El reconocimiento temprano de esta complicación, el manejo quirúrgico agresivo y el apoyo multidisciplinario son las bases para disminuir la morbimortalidad.

INTRODUCTION

HELLP syndrome is named for the clinical entities that comprise it: hemolysis, elevated liver enzyme levels, and low platelet counts forming the acronym. It was first described in 1982 and represents a severe form of preeclampsia, which biochemically is characterized by hemolysis, elevated liver enzymes, and low platelet counts.¹ It has a frequency of 0.17 to 0.85% of all pregnancies and its presentation is more

common in multi-gestation, Caucasian, and older women.² Reported complications secondary to this syndrome include the development of cerebral hemorrhage, renal failure, pulmonary edema, and hepatic rupture, all of which occur in 12.5 to 65% of all cases.³

Symptoms of a hepatic hematoma rupture in this syndrome are mainly the presence of persistent epigastric severe pain, or right upper quadrant abdominal pain, associated to hypovolemic shock.

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Termination of pregnancy in these cases should be expeditious.⁴

The formation and rupture of a subcapsular hematoma at the hepatic level represents one of the most serious events for the mother and the product. The morbimortality rate is extremely high, so most cases require timely, intensive, and multidisciplinary surgical management. The incidence of these complications varies between 1:40,000 to 1:250,000 pregnancies.⁵ Spontaneous rupture of the subcapsular hematoma can produce profuse intraperitoneal bleeding, endangering patient's life. This entity is associated with a maternal mortality of 60 to 80% and fetal mortality of 56 to 75%.⁶ In Mexico, 79 maternal deaths due to hepatic rupture were reported by Vitelio Velasco over a period of 15 years.

We present the case of a woman with HELLP syndrome, who presented with ruptured hepatic subcapsular hematoma and severe hypovolemic shock that was successfully treated at the *Hospital de Especialidades del Centro Médico Nacional La Raza*, Mexico.

CASE PRESENTATION

A 32-year-old woman with an obstetric history of three pregnancies, two products obtained by cesarean section, as well as a donor nephrectomy in 2011, presented in her third pregnancy at 34 weeks of an apparently normal gestation. She began suffering 48 hours prior to admission to our hospital with a clinical picture characterized by abdominal pain in the epigastric region; 24 hours after the onset of the clinical picture, nausea and vomiting were added with persistent elevation of blood pressure and the finding of thrombocytopenia, for which reason she was sent to a second level hospital where severe preeclampsia and HELLP syndrome were diagnosed.

On admission, she was found to have hypertension that was difficult to control with a mean arterial blood pressure ranging between 100 and 120 mmHg. The initial laboratory test reported were as follows: hemoglobin 10 g/dl, hematocrit 32%, with

schistocytes in the smear, platelets 99,000 mm³, AST 402 U/L, ALT 32 U/L, uric acid 4.8 mg/dl, alkaline phosphatase 108 U/L; urinalysis was reported as pathological, with a protein level of 400 mg/dl, and countless white blood cells.

An obstetric US was performed showing a single live product with a fetal heart rate of 140 beats per minute. An emergency cesarean section was performed, obtaining a single live product, and finding a subcapsular hepatic hematoma of approximately 10 × 10 cm, for which an exploratory laparotomy was performed in the same surgical event. Hemorrhage was estimated in 1,500 ml. A hepatic packing was placed, and she was subsequently admitted to the Intensive Care Unit. At that time, she was hemodynamically stable, with mean arterial pressure above 70 mmHg. Two hours later she developed hypovolemic shock, arterial hypotension, with evidence of bleeding, and showed a hemoglobin level of 5.5 g/dl and a platelet count of 80,000 mm³. A new surgical exploration was performed that revealed hemoperitoneum of 1,000 ml and hepatic hemorrhage at the level of the round ligament and at the intersection of both lobes. A new hepatic packing was placed. The reported bleeding was 1,000 ml during the procedure. The patient was readmitted again to the Intensive Care Unit, and up to this moment six red blood cell concentrates, 10 bags of fresh frozen plasma and six platelet apheresis had been transfused. She was sent to a third level of care hospital. At the moment of her arrival to this last hospital, she was found to be in a pretty bad condition, with mechanical ventilatory support, blood pressure of 89/58 mmHg and a hemoglobin level reported in arterial blood gases of 5 g/dl, as well as data of consumption coagulopathy. She was admitted directly to the operating room where a hemoperitoneum of 1,500 ml and a broken subcapsular hepatic hematoma in the left lobe grade III were found. Ligation of the common hepatic artery and accessory left hepatic artery was performed, as well as a new hepatic packing with four compresses was placed (*Figure 1*). Three

units of fresh plasma and five units of red blood cell concentrates were transfused in the intraoperative period.

The patient was admitted to the Intensive Care Unit in a stable condition with a mean arterial pressure of 70 mmHg. A hepatic unpacking was performed 48 hours later without evidence of active bleeding. The liver showed adequate consistency, color, turgor, and a limited hematoma (*Figure 2*). Twenty-four hours later, mechanical ventilatory support was withdrawn without complications. Thereafter, she had an adequate clinical evolution with diet tolerance, and drains without evidence of active bleeding. Laboratory tests taken after seven days showed a hemoglobin level of 11 g/dl, platelet count of 332,000/mm³, and coagulation times within normal parameters. Also, the liver function tests were found to be within normal parameters. She was discharged from the Intensive Care Unit in stable condition. Nine days postoperatively she presented persistent fever, and a CT scan was performed showing a large area of necrosis in the left hepatic lobe and an intraparenchymal hematoma of 15 × 10 cm (*Figure 3*), which lead to perform another exploratory laparotomy with the finding of a necrotic left hepatic lobe and an extensive hematoma of 1,500 ml (*Figure 4*); the hematoma was evacuated and a left hepatectomy was performed (*Figure 5*).

Subsequently, the evolution was favorable, and the patient was discharged 10 days later with a normal control CT scan, platelets within normal parameters, liver function tests without alterations, and proteinuria that had decreased to 50 mg/dl. The patient continued to be followed up by the nephrology service. She had complete recovery of renal function, and no subsequent events of high blood pressure.

DISCUSSION

Hepatic rupture in HELLP syndrome is a rare complication with high mortality, which compromises the life of the mother-child pair. Initially described by Abercrombie in 1844, it presents clinically with abdominal pain

predominantly in the right hypochondrium or epigastrium, right shoulder pain, nausea, vomiting and evidence of hypovolemic shock.⁴ Hepatic rupture can occur before, during or after delivery.

The cause of subcapsular and intrahepatic hematoma formation in HELLP syndrome is not yet well understood, but it has been shown that patients with eclampsia develop areas of necrosis and fibrin deposits in the hepatic sinusoids, causing obstruction to blood flow and hepatic distension, resulting in epigastric and right hypochondrium pain.⁷ The scar tissue in these cases is at greater risk of rupture due to the elevated blood pressure characteristic of this syndrome, forming a subcapsular hematoma, which tends to rupture easily due to any trauma or the expansion of the hematoma itself.⁶

Hematoma formation is more frequent in the right hepatic lobe. In a study by Henny and collaborators in which 75 patients with hepatic hematomas were included, it was found that in 75% of the cases the hematoma was in the right lobe, 11% in the left lobe and 14% in both lobes.⁷

Management of this complication requires clinical suspicion, as well as early diagnosis and aggressive treatment. The diagnosis should include, in addition to laboratory tests aimed at monitoring the alterations inherent to HELLP syndrome (blood cell count, and coagulation and liver function tests), imaging studies to help us determine the size of the hematoma and the amount of free intra-abdominal fluid. Ultrasound is the study of choice in most cases, but computed tomography (CT) scan has a great sensitivity to determine these alterations. However, it is not indicated as a first study in patients with hemodynamic instability.

Interruption of pregnancy is an important part of treatment as it eliminates placental stimulation, so pulmonary maturation should be initiated in patients with pregnancies of less than 34 weeks of gestation and severe preeclampsia, and blood pressure control is essential.⁴

Few reports have been described of non-surgical management in cases of small and contained hepatic hematomas, in

which the evolution of the hematoma must be strictly monitored by imaging control. This approach is only indicated in patients with hemodynamic stability and without coagulopathy.^{8,9} Surgery is the procedure of choice, since it is associated with better survival. The refinement of surgical techniques has led to a decrease in mortality, with a 30% reduction described in the latest published series.⁵

Surgical options described include hepatic packing, fibrin sealant management, direct suturing of damaged liver tissue, hepatic artery ligation or embolization, hepatic lobectomy, orthotopic liver transplantation, or a combination of these procedures.¹⁰ Patients should be treated in a center with expertise in liver trauma or by surgeons specializing in liver surgery and transplantation.

Hepatic packing by means of compresses does not always guarantee good results since the patients present with thrombocytopenia. It is especially important to perform a correct packing. The complete mobilization of the liver is achieved by means of the section of its ligaments. Care must be taken in cases with contained subcapsular hematoma, since avulsion of the hepatic capsule can be provoked.

Local control of hepatic hemorrhage, using hemostatic agents or suturing the bleeding

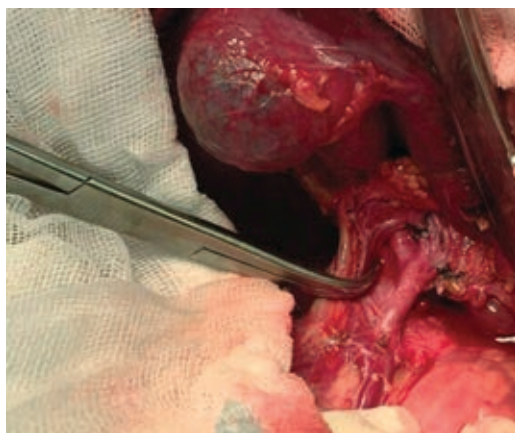


Figure 1: Dissection of the hepatic hilum with ligation of the common hepatic artery and accessory left hepatic artery.

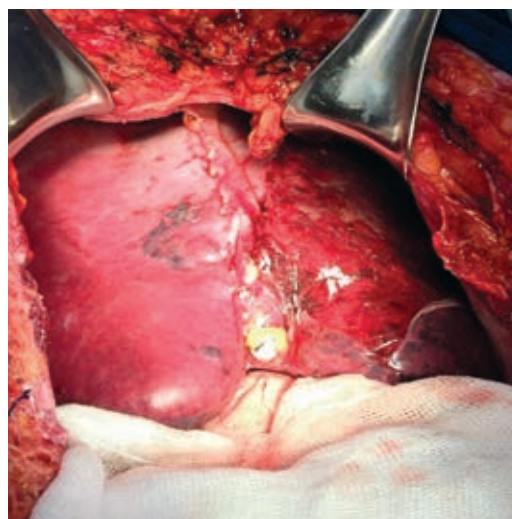


Figure 2: Appearance of the liver during unpacking (48 hours later), without evidence of active bleeding, with limited hematoma.

hepatic surface, is of limited value with a low success rate especially in cases of extensive hematomas.¹¹

Interruption of hepatic artery flow, either by direct ligation or angioembolization, has been used in various pathologies such as hepatic trauma, ruptured hematoma, or spontaneous hepatic hemorrhage, is in many cases an effective method of hemorrhage control.¹² In general, hepatic artery ligation is well tolerated. It is common to find transient elevation of liver enzymes, mainly aspartate aminotransferase (AST) and alanine aminotransferase (ALT), and it is important to remember that in patients with chronic hepatic pathology these enzymes may be already increased. Cholecystectomy is always necessary, since ligation proximal to the origin of the cystic artery may cause vesicular gangrene. It has been shown that the development of hepatic reperfusion through collateral arteries after hepatic artery ligation develops approximately 10 hours after the procedure;¹³ however, one of the drawbacks of this method is the development of focal areas of hepatic necrosis, which can become infected.

Arterial ligation can be performed selectively in the right hepatic artery, left

hepatic artery or common hepatic artery; this will depend largely on the characteristics and site of the hematoma. A transient clamping can be performed to determine if there is a decrease in bleeding, and then the definitive procedure can be performed by ligation,

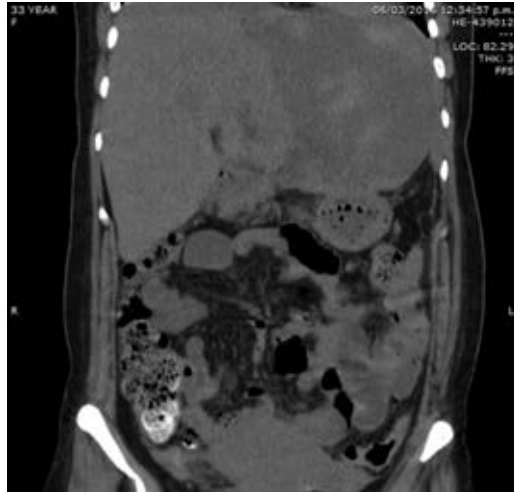


Figure 3: Tomographic control nine days after with findings of necrosis in the left hepatic lobe and an intraparenchymal hematoma measuring 15 × 10 cm.

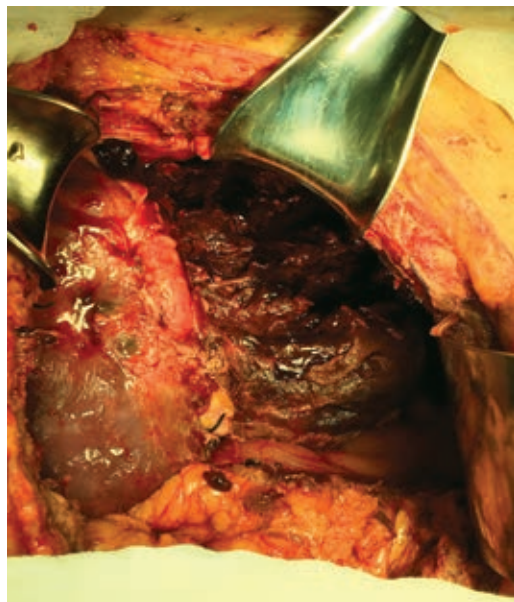


Figure 4: Exploratory laparotomy nine days after surgery with necrotic left hepatic lobe and an extensive hematoma of 1,500 ml.

making sure that there are no aberrant hepatic arteries. The left hepatic artery is generally a branch of the left gastric artery, and the origin of the right accessory hepatic artery may be at the level of the superior mesenteric artery.⁴ In the study performed by Araujo, ligation of the artery was reported in 50% of patients with hepatic hematoma, with good results in the control of hepatic hemorrhage and no evidence of subsequent liver damage.⁸

The most severe cases occur when there is an important degree of hepatic necrosis or devascularization. If these are limited, anatomic or non-anatomic hepatic resections can be performed, according to the area involved. In cases of uncontrollable bleeding or fulminant hepatitis, orthotopic liver transplantation is the treatment of choice, which should be performed before the appearance of septic complications or multiorgan failure.¹⁴

The treatment in each case must be initially focused on saving the patient's life, which sometimes implies performing

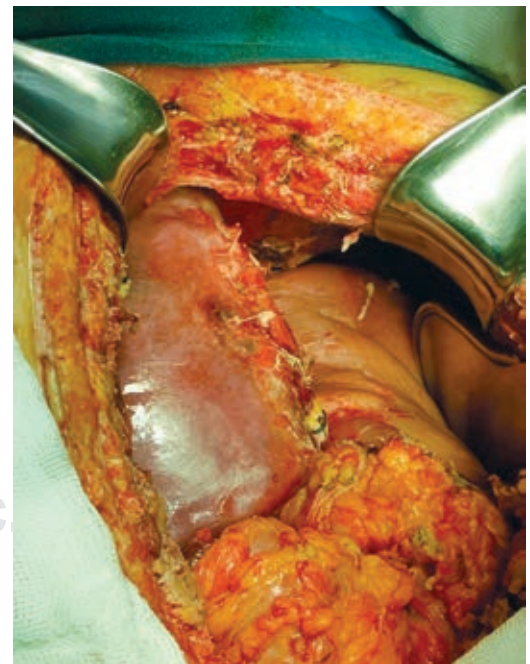


Figure 5: Appearance of the liver remnant after left hepatectomy.

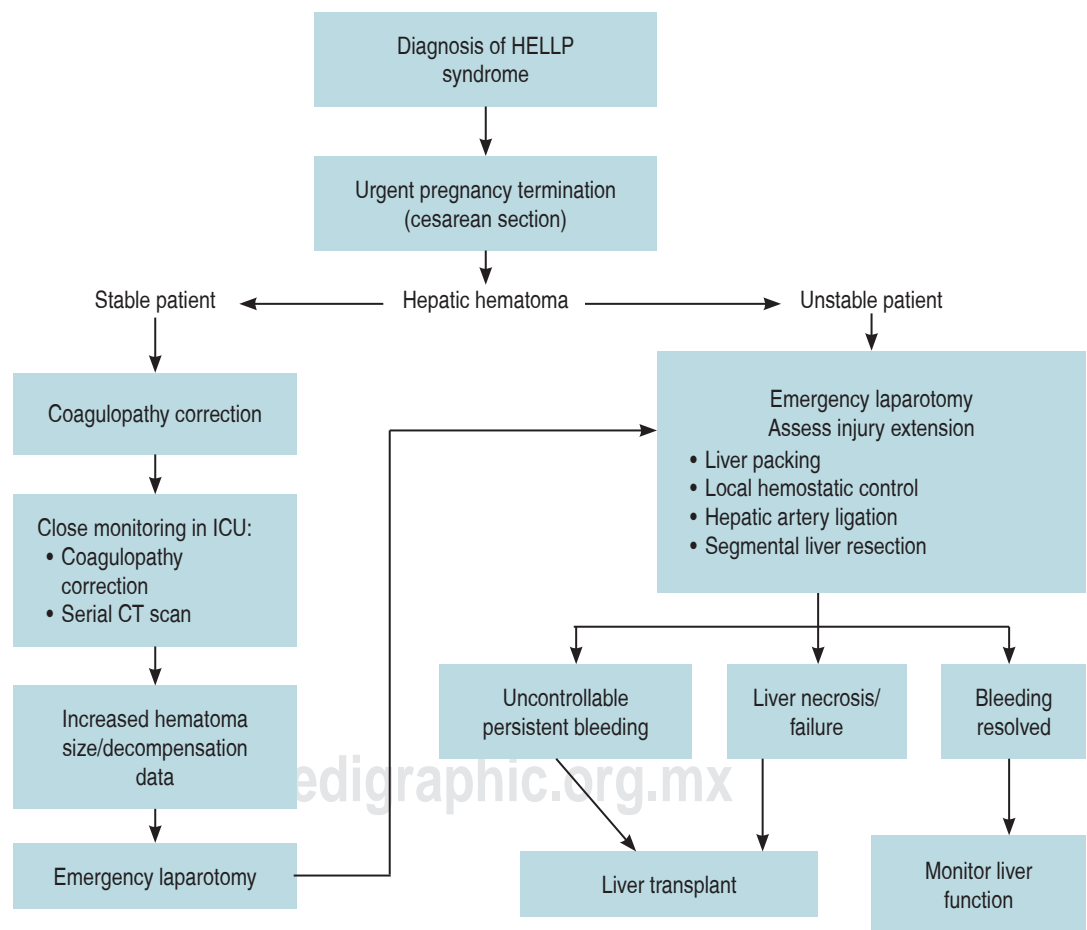
a first surgical intervention for damage control, with hepatic packing and arterial ligation, sending to intensive care to improve the patient's general conditions and, in a second stage, performing the unpacking and a second look surgery, in which the definitive treatment can be performed. Resolutive management may involve the performance of segmental liver resections or, in the most severe cases, liver transplantation (Figure 6).

HELLP syndrome is a late complication of preeclampsia-eclampsia, which makes it necessary to emphasize the importance of medical management as one of the most important preventive measures to avoid the occurrence of hepatic rupture, in addition to

the importance of a good pregnancy control, even when the pregnancy is of low risk, with monitoring of blood pressure, and laboratory tests that include blood cell counts, liver function tests and urinalysis, with the aim of detecting preeclampsia or eclampsia in a timely manner.

CONCLUSIONS

Hepatic rupture in HELLP syndrome is a serious complication that continues to have a high mortality rate worldwide. Early recognition of this complication together with aggressive surgical management and multidisciplinary support are the basis for reducing the associated morbidity and mortality. Multiple surgical



Transplant Department, HE CMN La Raza. 2017.

Figure 6: Management of hepatic hematoma in HELLP syndrome.

techniques have been described with different results, it is important to evaluate each case according to its severity and to refer to centers specialized in liver surgery and transplantation in a timely manner.

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Ethical considerations and responsibility:

Data privacy. In accordance with the protocols established at the authors' work center, the authors declare that they have followed the protocols on patient data privacy and preserved their anonymity. The informed consent of the patient referred to in the article is in the possession of the author.

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